Bladder extrophy "urinary diversion"

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Abstract

Introduction: The surgical treatment of bladder extrophy presents a major problem, particularly in delayed admitted cases and in those with a small bladder plate as well as failed cases of bladder extrophy closure.
Materials and Methods: In a study on fourteen out of the thirty cases of bladder extrophy, a rectosigmoid pouch was separated from the colon; the ureters were then implanted in the pouch with colon pull-through inside the reconstructed bladder ("rectosigmoid pouch") with complete separation of urine and stool. The operation was carried out in 3 stages.Of these 14 patients only seven underwent the 3 stages.
Result: In the follow-up study after 20-40 years, of the seven patients that underwent all three stages, all of them were enjoying an active life with satisfactory growth and development.
Conclusion: In the absence of any better options for the treatment of bladder extrophy, the above mentioned operative procedure can be recommended as it is safe and satisfactory with relatively good long term results.
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Keywords

- Bladder extrophy
- Bladder augmentation
- Colonic conduit
- Ileal conduit (Bricker procedure)
- Continent urinary diversion
- Ureterosigmoidostomy

Introduction

History

The earliest description of bladder extrophy can be seen on an Assyrian tablet from 2000 B.C. at the British Museum in London. Urinary diversion was attempted as early as 1851 in an extrophied patient. The first successful ureterosigmoidostomy was performed by Syme in 1852.[1] In 1892 Maydl transplanted the trigone into rectum.[2] In 1898 the construction of rectal pouch with transsphincteric colon pull-through was reported by Gersuny

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and Heintz Boyer in 1912.[3, 4] In 1909 Coffey performed ureter intestinal anastomosis with a long tunnel for entry of ureter into colon.[5] In l950 Bricker transplanted both ureters in isolated ileal segment with ileostomy ("ileal conduit").[6] In 1953 Mathisen reported the creation of an intraluminal ureteral nipple.[7] In 1954 Leadbetter and Clark described combined ureterosigmoidostomy.[8] In 1983 Hendren modified the ureterocolic anastomosis with better results.[9] In 1969 Kock reported a reconstructed continent ileal pouch with catheterizable intussucepted nipple after total coloproctectomy.[10] In 1973 the Mathisen ureterosigmoidostomy was applied for the first time at the University Clinic Children's Hospital Teheran, where a few years later, the Politano-Leadbetter ureterocystoneostomy was used for the reimplantation of ureters in the colon and reconstructed pouch. Anatomic reconstructions of the bony pelvis in extrophied patients were first attempted by Trendelenburg in 1906 ("bilateral posterior sagittal iliac osteotomies").[11] In the last decades supraacetabular pelvic osteotomy was used very often. The combined anterior innominate and posterior vertical osteotomies for the anatomic reconstruction was reported by Gearhart and Associates.[12]

The surgical procedure ("Continent rectal bladder reconstruction for urinary diversion with complete separation of stool and urine by colon pull-through inside the reconstructed bladder") was performed by the author since 1976 at the Children's Hospital Medical Centre Tehran.

General remarks

The surgical treatment of bladder extrophy presents a major problem, particularly in delayed admitted cases and in those with a small bladder plate. An increasing number of failed cases of bladder extrophy closure in neonates and infants in the last decades present a further major problem area for surgical treatment. The ideal treatment of this congenital anomaly is yet to be established despite various available options.

About forty four years ago (mid 1970's) a rectosigmoid pouch was separated from the colon, the ureters were then implanted in the pouch, followed by coloproctostomy at the Children's Hospital Medical Center Tehran. This operative method was presented for the first time by the author at the 4th World Symposium of Pediatric Surgery in Barcelona July 26-29, 1977. The disadvantage of this operative technique was the incomplete separation of urine and stool. Four years later, the author modified this operative technique with colon pull-through inside the reconstructed bladder ("rectosigmoid pouch") with complete separation of urine and stool. This operative technique is not indicated in children with weak anal tone and contraction and should be carried out in patients with a perfectly functional anal sphincter. The operative procedure is performed preferably in three stages, usually when the child is older than 2-3 years. Without stoma, appliance or catheterization, this type of continent urinary diversion is especially suitable for children. It is a relatively safe and good alternative to other continent urinary diversions.

Materials and Methods

Operative procedure

Stage I

Preoperative treatment and preparation

Five days prior to the operation oral metronidazol is given. Forty-eight hours before the Operation, rectal wash-out is conducted 3 times daily with normal saline, in which 1-2 Dulcolax tablets have been dissolved each time and repeated several times if needed. A broad spectrum antibiotic is started intravenously on the day of the operation.

Positioning of the child and skin incision

The child is placed in a supine position. A longitudinal skin incision in the lower abdomen is conducted which continues semi-circumferentially to the left of the umbilicus.

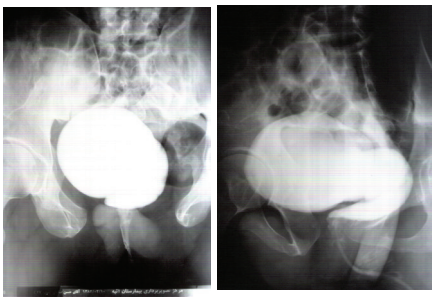

Figure1: Ascending Pouchogramme “Rectosigmoid Pouch”
25 years post-op, reconstructed bladder and urethra.

Approach and operative technique

The Trendelenburg position is applied for median laparotomy in the lower abdomen. A retractor is inserted and the small bowel loops are pushed back with moist lap pads and Deaver retractor. The descending colon and rectosigmoid colon are exposed. The inferior mesenteric artery with the branches to the rectosigmoid colon is exposed. The rectosigmoid colon is transected about 20 cm above the peritoneal reflection and the mesocolon with the supplying vessels to the separated rectosigmoid colon is divided **Figure 2.**

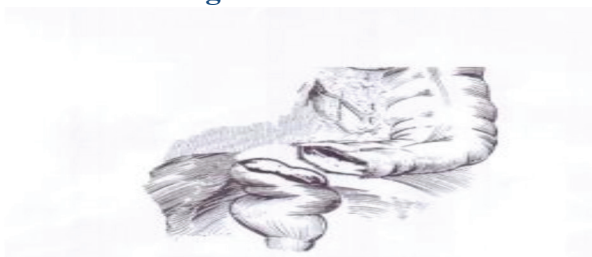

Figure 2: Transection of the rectosigmoid colon

The proximal colonic segment is mobilized with an incision of the peritoneum lateral to the colon and division of the branches of the sigmoid mesenteric vessels. Care should be given to the vascular supply of the rectosigmoid colon and to the supplying vascular arcade of the proximal colonic segment. The end of the proximal colonic segment is reanastomosed end-to-side to the anterolateral wall of the rectum a few centimeters above the peritoneal reflection using interrupted 2-0 sutures. The distal segment is detubularized by anterior opening in the proximal half along the tenia **Figure 3**.

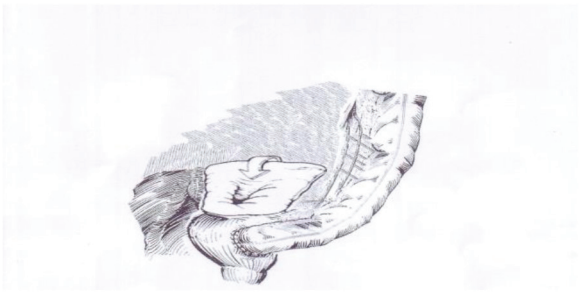

Figure 3: Anterior opening of the distal colonic segment

Two submucosal tunnels are created on the rectosigmoid plate. The peritoneum is incised bilaterally and the ureters mobilized retroperitoneally for a tension free anastomosis. A seromuscular incision just above the submucosal tunnel is followed by antireflux ureteral implantation. The first 3 sutures for the ureteral anastomosis are done with chromic 4/0, taking the colonic mucosa with the muscular layer and ureter in the lower part. The remaining stitches take only the mucosa and the ureter. Two feeding tubes are inserted in the ureters and they are taken exteriorly through the anus **Figure 4**.

Figure4: Creation of 2 submucosal tunnels

The rectosigmoid plate is folded anteriorly on itself and closed with vicryl 2/0 running sutures to form the rectosigmoid pouch. Lateral adaptation stitches between the pouch above the entrance of the ureters and the peritoneum protect the ureter and the anastomosis. A rectal tube is inserted through the colorectal anastomosis in the colon **Figure 5.**

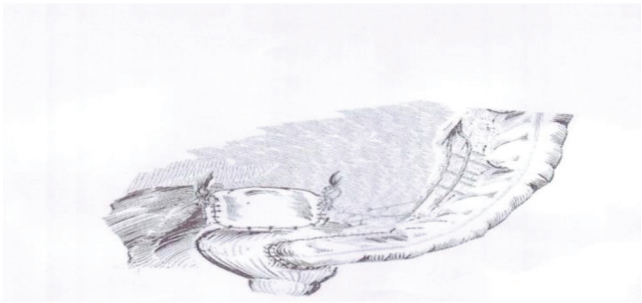

Figure 5: Closure of the rectosigmoid plate

Post-operative treatment

Intravenous alimentation is maintained until normal bowel function resumes, usually for 5-6 days. The rectal tube is usually removed after 3-4 days, and the ureteral stent after 5-6 days. Intravenous broad spectrum antibiotics and metronidazole are continued for a week and thereafter, prophylactic per oral therapy for another 7-10 days. Follow-up study is carried out using: ultrasound, DTPA-renal scan and blood gas analysis.Also ascending pouchogram 4 weeks before the second stage operation is done to rule out reflux.

Stage II

Preoperative treatment and preparation

This is done the same as in the first stage.

Positioning of the child and skin incision

This is done the same as in the first stage.

Approach and operative technique

The second stage operation is usually performed

after a year. The rectal bladder is enlarged during this time, mostly with a capacity of more than 300-400 ml and it is almost spheric in shape. Median laparotomy is performed in the lower abdomen, a retractor is brought into the wound, and the rectal pouch in the pelvic cavity is prominent. The adhesion of the pouch to the surrounding structures is dissected free and the colorectal end-to-side anastomosis is exposed. A vertical incision is performed with diathermy in the middle on the anterior surface of the rectal pouch from the dome to the colorectal end-to-side anastomosis and the distal colon is separated from the rectal pouch on the anastomosis. The ureteral orifices are catheterized bilaterally **Figure 6.**

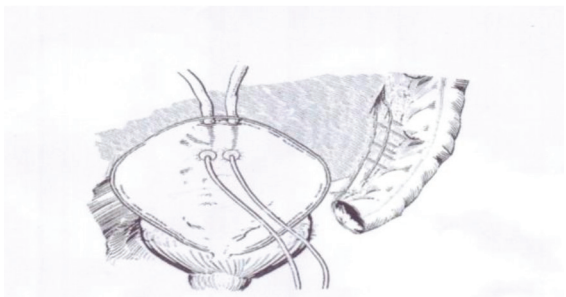

Figure 6: Opening of the rectal pouch anteriorly with separation of colorectal anastomosis

If there is stenosis or reflux, they should be corrected. The opened pouch should be taken bilaterally with 3 to 4 stay sutures. The mucosa on both sides is carefully dissected free from the pouch wall with a blunt Stephen’s scissors up to the middle of the pouch, so that the proximal colonic segment can be pulled through in the anterior part of the pouch with ease **Figure 7**.

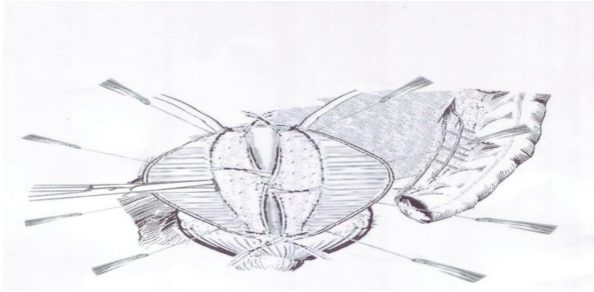

Figure 7: Bilateral Dissection of the pouch mucosa free up to the middle of the pouch

Care should be taken not to damage the rectal mucosa. If there is a hole in the mucosa, it should be repaired with chromic suture. The dissection of the mucosa on both sides is continued further downwards to the anal canal and the ureteral stents are taken out through the anus. The mucosa is approximated on both sides and closed in the middle with vicryl 5/0 running sutures **Figure 8**.

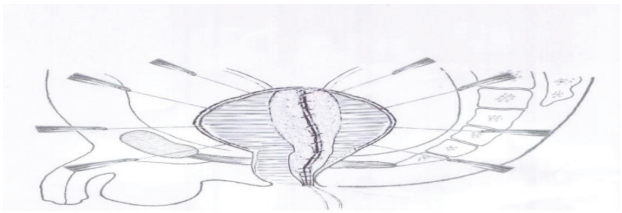

Figure 8: Closure of the mucosa in the middle of the pouch and extension to the anal canal

The child is then brought to a lithotomy position and the anus is exposed and opened with 2 small blade Langenbeck retractors. The lower part of the mucosa in the anal canal is dissected free bilaterally and closed in the midline with interrupted 5/0 vicryl single sutures over the ureteral stents **Figure 9**.

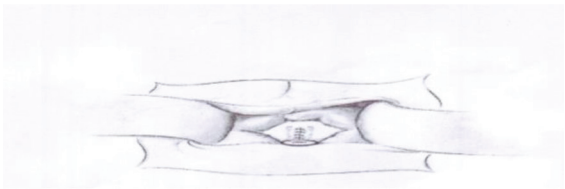

Figure 9: Closure of the mucosa in the anal canal on the posterior surface

Two small mucocutanous flaps are taken on the posterior aspect of the anus bilaterally, overlapping the distal end of the reconstructed neourethra over the ureteral stents. Thereafter, the distal colonic segment in the lower abdomen is tapered in the terminal part for a distance of 5-6 cm and pulled-through in the anterior part of the rectal pouch over the joining mucosa to the anus. The tapered part of the terminal colon should be facing anteriorly to the anus **Figure 10**.

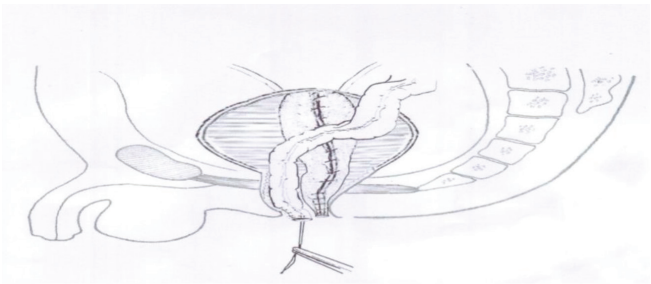

Figure 10: Pull through of the proximal colonic segment

The anastomosis is performed with vicryl 3/0 or 2/0 interrupted single sutures between the end of the proximal colonic segment and three quarters of the anterolateral circumference of the anus. A rectal tube is inserted in the colon **Figure 11**.

Figure 11: Anastomosis between the end of the proximal colonic segment and three quarters of the anterolateral circumference of the anus

The seromuscular wall of the pouch is approximated in the middle over the colon and closed with vicryl 2/0, and fixation of the entrance of the colon to the pouch is performed with 2/0 interrupted single sutures between the seromuscular layer of the colon and the wall of the pouch **Figure 12**.

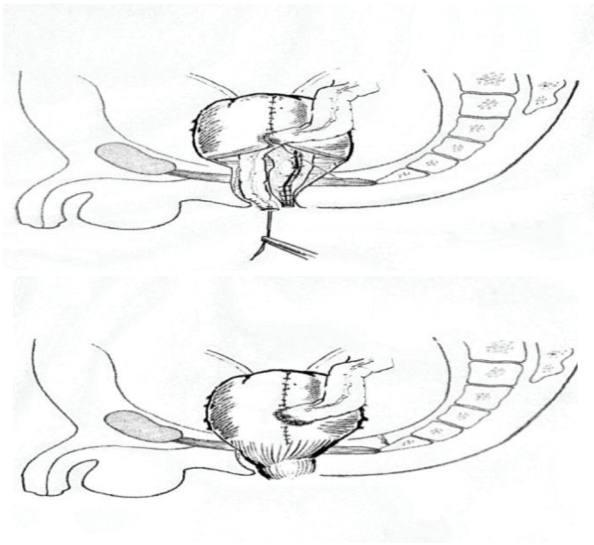

Figure 12: Closure of the seromuscular wall of the pouch

Postoperative treatment

This is the same as in the first stage.

Stage III

This is usually performed 1-2 months after the second stage operation. Supra-acetabular pelvic

osteotomy is performed for approximation of the diastasis pubis and the external genitalia are reconstructed.

Results

Between 1973-1982, thirty cases of classic bladder extrophy in infants from 4 months to 5 years of age were surgically treated by the author at the University Children’s Hospital Medical Center in Teheran. The ratio of boys to girls was 3/1. During this time, a rare case of incomplete bladder extrophy was also observed and surgically treated.[13]

The operation described above was performed in fourteen out of the thirty cases. However, during 1973-1982, the three stages of the surgical procedure could be completed in only two of the cases. For the remaining twelve cases, only the first stage of the operation was performed during 1973-1982. Since 1992, five of the patients came for the second and third stage of the operation. Thus, altogether seven patients out of the fourteen completed all three operative stages. During 1973-1982 ureterosigmoidostomy and primary closure were performed in sixteen other cases. In the follow-up study after 20-40 years, of the seven patients that underwent all three stages, all of them were enjoying an active life with satisfactory growth and development. In one of these patients, 14 years after first stage operation, there was ureter obstruction on both sides due to UVI stenosis with renal damage, which could have been corrected during the second stage operation. In another case, after 16 years, VUR was observed on both sides, which was also corrected by ureter reimplantation. In another patient, after 36 years, colon obstruction occurred due to stenosis of the colon on the entrance to the reconstructed bladder; and was surgically corrected. Signs and symptoms of UTI in these patients are urinary frequency, lower abdominal discomfort, feeling ill, inappetence, fatigue, fever, flank pain and sometimes diarrhea. Since urine culture is not significant in such cases, the antimicrobial treatment should be started immediately. In case of recurrent UTI, it should be fully investigated. After separation of the reconstructed bladder and rectum, signs and symptoms of UTI were rare in our patients, so long as it was not caused by reflux or UVI stenosis, and disappeared after a day or two with anti-microbial treatment. Since most of these patients lived a long distance away from Tehran, it was difficult to do regular annual checkups. In some cases they came for a checkup after five to ten years. We therefore gave them instructions to start immediate treatment as soon as signs and symptoms of UTI appeared.

The most intensive renal damage among our cases occurred between the first and second operative stage during a delayed interval of 14 to 20 years. During this time, the urine and feces were mixed in the rectosigmoid pouch and colon. Therefore the risk of pyelonephritis was relatively high. After completion of the second and third stage operation, there was a complete separation of the reconstructed bladder and rectum, lasting twenty to twenty four years. The comparison of DMSA renal scan and renal sonography between the two periods, prior to and following separation, revealed that the major renal tissue damage was caused by urine and feces admixture. The latest laboratory examinations revealed acceptable BUN and creatinine in three of the patients, but had increased in four patients. Nonetheless, all of these patients were still in an

acceptable condition and were enjoying an active life. All these patients were voiding spontaneously, in five cases with a dry interval of three to four hours and more. In two cases, there was a dry interval of two to three hours. All of them were continent for stool during day and night. We were able to do a urodynamic study in three cases, all of which showed capacious and low pressure reconstructed bladder.

Regular follow-up is indicated once a year with CBC, arterial blood gas analysis, ultrasound, DTPA-renal scan.If there is sign of recurrent UTI, or worsening of the renal function, DMSA renal scan and ascending pouchogram from the reconstructed bladder reservoir to roll out obstruction or reflux should be carried out. Endoscopy of rectal bladder with mucosa biopsy, and urodynamic study is indicated between 5-10 years after the operation. Further follow-up studies concerning management, different operative methods and results, as well as complications will be presented in the discussion.

Discussion

Ureterosigmoidostomy

Conventional ureterosigmoidostomy has often been used as the primary option for urinary diversion. It is an easy procedure and the use of external urinary devices, catheterization and stoma is not necessary. Yet this procedure has its own major complications such as a high incidence of urinary proctitis, electrolyte imbalances, metabolic acidosis, hyperkaliamia, bone demineralisation, growth retardation, hydronephrosis, hydroureter, ascending pyelonephritis, renal calculi, uremia, and development of neoplasma of the colon.

Primary reconstruction

The surgical reconstruction of bladder extrophy is one of the most challenging efforts in pediatric surgery. The standard treatment of most cases of bladder extrophy epispadias complex at present is primary bladder closure along with bladder neck reconstruction, osteotomy and also anti reflux surgery. The ultimate purpose of surgical treatment is for the patient to have continence, maintain a healthy upper urinary tract and create an adequate genitalia and body image. The achievement of balance between intravesical pressure and outlet resistance in cases of primary bladder extrophy closure with bladder neck reconstruction is difficult in the majority of cases, often leading to secondary procedures due to renal damage and poor continence. A study by Ransley and Associates on bladder function and dysfunction in children with extrophy and epispadias, show sever dysfunction of bladder after conventional bladder neck surgery for continence.[14,15] These authors state that despite what is believed, normal function is not achieved in children with closed extrophy bladders and this is a major cause of upper-urinary-tract damages and could harm the development of bladder capacity. They also state that 25-50% of children with bladder extrophy who are born with healthy kidneys, develop upper-urinary-tract damage after reconstruction procedures. They also found that a common complication of bladder neck reconstruction is an inability to contract the detrusor which is the main reason for leakage due to bladder instability. Another urodynamic study performed by Hendren, Retik and Associates shows that following bladder neck reconstruction compliance and stability are impared.[16] About 1in 4 cases of bladder extrophy may still have normal detrusor function

following reconstruction. Other studies report the same problem and results.[17,18] They concluded that continence is not always achieved following a successful surgery of bladder extrophy. Some patients, who in initial assessments appear to be great candidates for successful continence surgeries, may experience failure of the procedures and thus needing future bladder augmentation or urinary diversion. Immunohistochemically and morphometric studies of extrophic bladders at different stages of reconstruction have shown differences in the type of collagen and smooth muscle-to-collagen ratio. Also, the newborn extrophic bladder seems to have less myelinated nerve fibers.[19, 20]

Bladder augmentation

Bladder augmentation is another alternative for the surgical treatment of bladder extrophy. However, this treatment is accompanied by a large number of complications, such as mucous production and urinary drainage problems, hematuria, bladder stones, bladder perforation, UTI, and pyelonephritis, renal stones, vesicoureteral reflux recurrences, ureterovesical junction stenosis, bowel obstruction, vesicourethral fistula, metabolic problems and malignant changes. When the absorptive intestinal mucosa is exposed to urine multiple metabolic and electrolyte abnormalities, including metabolic acidosis, occur. Of the effects of metabolic acidosis on the child’s body is bone demineralization and decreased bone growth. This is why in cases with a history of ileal ureters, ureterosigmoidostomy, and colocystoplasty; osteomalacia and osteopenic changes are seen.[21,22,23] Growth retardation and development is also reported in cases with bladder augmentation due to metabolic acidosis.[24] Another major complication in patients who have had bladder augmentation is bladder stone which is a major burden on the patient and the healthcare system.[25,26,27] The combination of bladder augmentation and Mitrofanoff is indicated in patients with difficult urethral catheterization.[28,29,30,3]

Continent urinary diversion

Another alternative for the surgical treatment of bladder extrophy is continent urinary diversion. During the past decade, there has been an increased interest in continent urinary diversion with better management of metabolic acidosis and the ability to create an antireflux ureteral-colonic anastomosis with continence provided by the patients’ own sphincteric muscles. Thus ureterosigmoidostomy has enjoyed somewhat of a revival. A number of investigators regard ureterosigmoidostomy as the preferred method of management in children with extrophy of the bladder if primary reconstruction is not possible.[32, 33, 34]

In the last 20 years there have been reports about diversion procedures which can prevent major renal losses and reduce problems due to urine collecting appliances. This approach was created by the continent urinary diversion technique (in which a partially separated capacious urinary pouch with low pressure is created that functions better after cystectomy).[35,36,37] In the follow-up this diversion procedure was further modified by Hohenfellner and associates by augmented valved rectosigmoid (“ileocecal ureterosigmoidostomy”).[38,39] Later, Retik reported a similar operative technique (“ureteroileocecal sigmoidostomy”) as continent urinary diversion.[40] The advantages of this procedure is that there is no anastomosis of the ureter to the

sigmoid and no urine-feces admixture at the site of ureteroileal anastomosis. The intussuscepted ileocecal valve protects the ureteroileal anastomosis from contact with feces.

Regarding the author's procedure, which includes the advantages of augmented valved rectosigmoidostomy, there are also other comparative advantages. The high urinary capacity of the rectosigmoid bladder with low pressure, provide good protection in relation to upper urinary tract and renal damage. There is usually complete separation of stool and urine and the resorption ability in the rectum is very low, so that hyperchloremic metabolic acidosis is not a major problem and alkalization therapy is not needed in most of our patients.

Another advantage of our procedure is the lack of occurrence of adenocarcinoma which is a major complication of ureterosigmoidostomy in the long term follow-up. The tumor occurs most often at the site of implantation of ureter into the sigmoid colon.[41] Albeit, there have been reports of cancer in the ileal or colonic conduit, bladder augmentation or continent urinary diversion. The average time of tumor occurrence after an operation is 25 years with a mortality rate of approximately 50%.[42,43] It was believed that the cancerogen was probably formed by the conversion of urinary nitrates to nitroamines by fecal bacteria.[44] Therefore all efforts should be made to avoid urine-feces admixture and to control infection. The author's procedure reduces these risks considerably due to the separation of stool and urine, whereas in the Hohenfellner and Retik procedure, the urine and feces are mixed in the rectosigmoid and colon. A 20-38 year follow-up of our patients with endoscopy of reconstructed bladder and mucosa biopsy has not so far revealed any cases of dysplasia or malignancy. In addition, like other continent urinary diversions, our procedure requires no external appliances, colostomy or catheterization, which may influence the social and psychological well-being of the patient.[45] Most of our patients are enjoying an acceptable and active life, which is especially important in children.

The construction of rectal pouch with transsphincteric colon pull-through reported by Gersuny or by Heitz-boyer Hovelaque in the past is accompanied by a high risk of anal sphincter damage and the possibility of stool and urine incontinence. Obstruction of distal colon due to ischemia with a need for colostomy has also been reported. In the author's technique, the colon is pulled through inside the reservoir in the anterior part. There is no risk of damaging the nerves and the anal sphincter, or the blood supply of the colon. The urethra in the anal canal is constructed with mucosa, and the posterior rectal wall provides a good protection for the reconstructed urethra.

Pelvic osteotomy with joining of the pubis bone is mandatory. This corrects the body image and walking. It is also very useful for the appropriate genital correction as well as for a better achievement of continence. Our cases with correction of diastasis pubis after pelvic osteotomy are continent for stool and urine during day and night.

Conclusion

In conclusion, in the absence of any better options for the treatment of bladder extrophy, the above mentioned operative procedure can be recommended as it is safe and satisfactory with relatively good long term results.

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